Abnormal Neck Veins

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A woman in her 50s was admitted after several months of progressive dyspnea on exertion and severe lower extremity edema. She had undergone aortic valve replacement for aortic stenosis, 2-vessel coronary artery bypass, and permanent pacemaker placement 1 year before presentation. She had a history of mantle chest radiotherapy for Hodgkin lymphoma at age 17 years, which was curative. After successful intravenous diuresis, neck examination at approximately 60° (Figure 1A and Video) revealed a regular carotid pulsation and discernible internal jugular venous pulse. After examination of the neck at rest, the patient took a small breath and held it for a few seconds. Hemodynamic data at left and right heart catheterization are shown in Figure 1B.

WHAT IS YOUR DIAGNOSIS?

A. Tamponade
B. Myocardial restriction
C. Right ventricular failure
D. Wide-open tricuspid regurgitation
Diagnosis
B. Myocardial restriction

Discussion
When examining the venous pulsation (Video and Figure 1A), one notes a clear a wave just before the carotid systolic pulsation, followed by a visible x descent and a noticeable v wave coincident with the end of carotid pulsation. The absence of a large c-v wave abolishing the x descent (right atrial ventricularization) suggests that severe (wide-open) tricuspid regurgitation is not present. Immediately after the carotid systolic pulsation, there is an abrupt disappearance of the venous pulsations representing a rapid or deep y wave descent (Figure 2A). A rapid y descent is suggestive of a constrictive or restrictive process and coincides with the beginning of the “square root sign” (rapid increase in right and left ventricular diastolic pressure immediately after systole [Figure 2B, arrowhead]) during right-left heart catheterization. Conversely, tamponade is associated with a blunted y descent reflecting continuous elevation of diastolic pressures, without significant phasic variations. A prominent Kussmaul sign was then observed on inspiration (Video and Figure 2B, arrowhead), but this sign is nonspecific and could be seen in all 4 proposed diagnoses since it represents elevated right-sided pressures (irrespective of cause) that increase with inspiration as blood rushes into the right-sided chambers. The differentiation between constrictive and restrictive physiologic states cannot be entirely made by isolated neck vein observation, since both processes will likely exhibit jugular venous distention, rapid descent, and Kussmaul sign as in our patient. Trans-thoracic echocardiogram demonstrated normal left and right ventricular size and function, evidence of increased left-ventricular filling pressure (medial E/e’ ratio, 30), moderate to severe tricuspid regurgitation (possibly related to pacemaker wire), estimated right ventricular systolic pressure of 34 mm Hg, normal aortic valve tissue prosthesis, borderline-dilated inferior vena cava size with normal inspiratory collapse, and no evidence of constrictive physiology or pericardial effusion. Right-left heart catheterization with simultaneous high-fidelity micromanometer recording of the left ventricular pressure tracing and right atrial pressure tracing (Figure 1B and Figure 2A) depicted the physical examination findings (a wave, x descent, v wave, and deep y descent). Right and left ventricular pressure tracings showed restrictive filling pressures and absence of respiratory enhanced ventricular interdependence (Figure 2B), confirming absence of constriction.

Hodgkin lymphoma is a curable cancer for most patients owing to advances in radiotherapy and chemotherapy. However, survivors of Hodgkin lymphoma are at a substantially increased risk of developing radiation-related cardiovascular complications throughout their lives.1 Radiotherapy for Hodgkin lymphoma has been associated2 with coronary, myocardial, pericardial, and valvular heart disease due to tissue fibrosis. Risk factors for radiation-induced cardiotoxicity include radiation dose greater than 30 to 35 Gy, greater field size, younger age at exposure, longer time since exposure, and concomitant cytotoxic chemotherapy.2,3 As such, history of chest irradiation in a patient presenting with heart failure symptoms should prompt evaluation of coronary, myocardial, pericardial, and valvular heart disease as well as pulmonary disease (our patient had moderate restrictive lung disease demonstrated by pulmonary function tests). Of these conditions, the differentiation between restrictive cardiomyopathy and constrictive pericarditis can be challenging but invaluable. Constrictive pericarditis may be curable by pericardiectomy, whereas restrictive cardiomyopathy is not and thus is managed medically.

Trans-thoracic echocardiography is the initial noninvasive diagnostic test of choice when evaluating patients for restrictive cardiomyopathy and/or constrictive pericarditis. Various criteria exist in differentiating constrictive pericarditis from restrictive cardiomyopathy by echocardiography.4 However, in a subset of patients, echocardiography may not be able to distinguish restrictive cardiomyopathy from constrictive pericarditis and hemodynamic catheterization may be required.5

A follow-up echocardiogram 1 year later showed increasing pulmonary hypertension, mild mitral stenosis, untrisitucip regurgitation, and a normally functioning aortic valve prosthesis.

Additional Contributions: We thank the patient for granting permission to publish this information.

REFERENCES